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A Typical Case of Classic Potter's Syndrome: A Case Report

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Abstract

Potter syndrome is a rare congenital malformation that primarily affects male fetuses; it is characterized by pulmonary hypoplasia, skeletal malformation, and kidney abnormalities. The pressure of the uterine wall due to oligohydramnios leads to an unusual facial appearance, abnormal limbs in abnormal positions, or contractures. The fetus generally dies soon after birth due to respiratory insufficiency. The baby was a live preterm male, born to a 30-year-old multigravida, out of a non-consanguineous marriage via cesarean section. There was no liquor at the time of delivery. The baby did not cry immediately after birth and required resuscitation, followed by mechanical ventilation. Multiple congenital anomalies suggestive of Potter's syndrome were noted including facial features, flattened nose, low protruding ear, retrognathism, and epicanthal folds with unilateral atresia of the choana. Chest X-ray showed small volume lung fields suggestive of pulmonary hypoplasia, and we had on ultrasonography bilateral polycystic kidney disease on ultrasonography. At 42 hours of life, the baby developed tachypnea and severe chest retractions and died due to respiratory insufficiency. Our case highlights the importance of regular prenatal checks and examinations in each pregnancy, which helps to collect suspected cases and improve knowledge of this syndrome for better management.

Keywords

Potter Syndrome, Pulmonary Hypoplasia, Potter's Facies, Polycystic Kidney, Oligohydramnios

1. Introduction

Potter syndrome is a rare clinical syndrome first described by Edith Potter in 1946 at the Chicago Hospital in the USA. Characterized by facial features in infants with bilateral renal agenesis and oligohydramnios [1]. Its incidence varies from 1 in 2000 to 5000, with an average of 1 in 4000, reported in 0.2% - 0.4% of autopsies of newborns who died immediately after birth. It is a rare complication of oligohydramnios, mainly affecting male fetuses and characterized by pulmonary hypoplasia and renal anomalies [2].

We report a case collected in the Department of Neonatology Intensive Care Unit.

2. Case Presentation

This is a male newborn from a non-consanguineous marriage, premature at 33 weeks of amenorrhea with antenatal diagnosis of polycystic kidney disease. The newborn was born to a 30-year-old mother, G3P2E2 (history of IUFD, one healthy 8-year-old child) with a poorly monitored pregnancy, delivered by Caesarean section on oligohydramnios with an Apgar at birth of 5/10 then 8/10 and a negative infectious anamnesis.

At birth, the newborn was conscious, hypotonic, hypo-reactive, weak sucking reflex, weak archaic reflexes. Hemodynamically stable, respiratory unstable with a Silverman score of 3/10 (marked subcostal indrawing with suprasternal indrawing and flapping of the wings of the nose) (**Figure 1(a)**), SaO_2: 85% in ambient air and 93 on 3 L of oxygen. Physical examination revealed a dysmorphic facies (potter's facies): flattened nose, Low-set ears, retrognatism and epicanthal folds with unilateral choanal atresia. Distended abdomen, presence of a mass occupying the right flank with limb deformities (bowing of legs) (**Figure 1(b)**). The rest of the examination was unremarkable.

Radiologically, a chest X-ray showed bilateral pulmonary hypoplasia (**Figure 2**); an abdominopelvic ultrasound was consistent with bilateral polycystic kidney disease (**Figure 3(a)** and **Figure 3(b)**).

(a) (b)

Figure 1. (a) Image showing the dysmorphic potter's facies in our patient; (b) the image showing distended abdomen with limb deformities (bowing of legs).

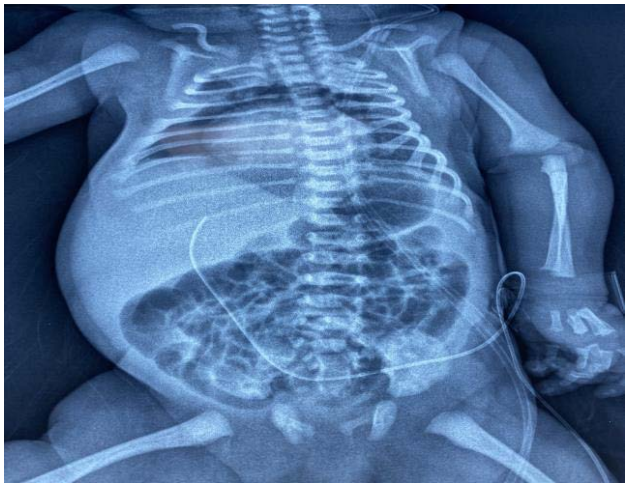
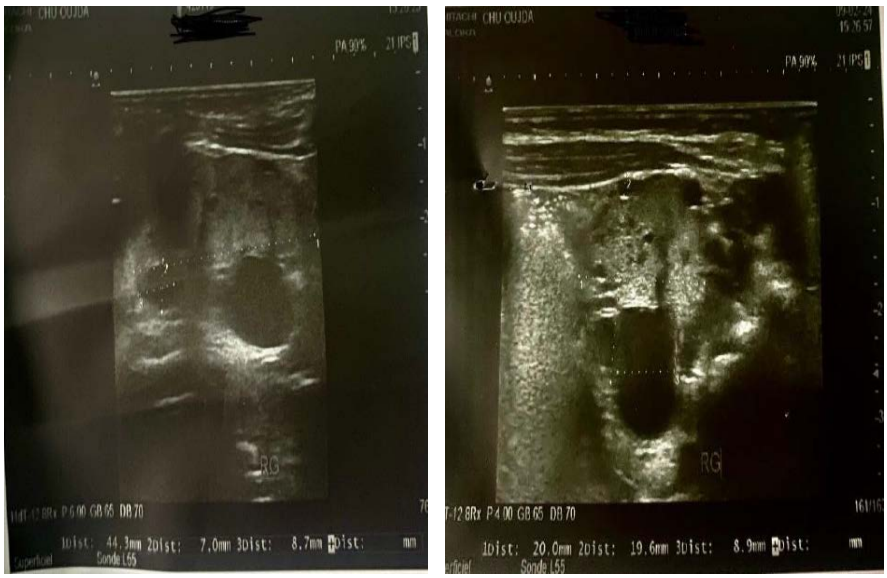


Figure 2. Chest X-ray showing bilateral pulmonary hypoplasia.



(a) (b)

Figure 3. (a) Ultrasound sections of the left kidney showing multiple cysts in our patient; (b) ultrasound sections of the left kidney showing multiple cysts in our patient.

The diagnosis of potter's syndrome had retained in view of the oligohydramnios, dysmorphic syndrome, pulmonary hypoplasia and polycystic kidney disease. The evolution had marked by worsening respiratory distress followed by cardiopulmonary arrest, which did not recover after well-conducted resuscitation measures.

3. Discussion

Potter syndrome described by Edith Potter in 1946, in newborns with bilateral renal agenesis or other renal anomalies, including aplasia, dysplasia, hypoplasia or polycystic kidney disease [3] [4].

Initially, the term was applied to cases presenting with bilateral renal aplasia

(Potter sequence), but today the term refers to an atypical morphological appearance of the newborn due to an underlying cause of oligohydramnios (fetal growth retardation, premature rupture of membranes, fetal or post-maturity chromosomal abnormalities, etc.). It can be classified into different types, the causes being renal and non-renal [5] [6].

The Potter sequence is thought to result from intrauterine compression of the growing fetus due to severe oligohydramnios leading to physical deformities, being the "Potter facies" [3] [5]. The latter is characterized by low-set ears, hypertelorism, retrognatism, widening and flattening of the nasal root, a parrot-beak nose and an epicanthus. Other features of the Potter sequence include pulmonary hypoplasia (the degree of pulmonary hypoplasia depends on the degree and duration of oligohydramnios, as well as the stage of lung development at which oligohydramnios occurs) [6]. Limb deformities (clubfoot, leg tilt, limb hypoplasia, etc.); ophthalmological malformations (cataract, lens prolapse, angiomatosis malformation of the optic disc area, etc.) VACTERL (vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal anomalies, limb anomalies), caudal dysgenesis, caudal dysplasia syndrome, and isolated anomalies of the skeletal and central nervous system [7] [8]. Skeletal abnormalities frequently associated with this condition are hemi-vertebral and sacral agenesis. These anomalies may aggravate the increased morbidity and mortality in these infants.

Normally during fetal development, fetus continuously swallows the amniotic fluid, which after getting reabsorbed by the gastro-intestinal tract, is again reintroduced into the amniotic cavity by fetal kidneys in the form of fetal urine. If the volume of amniotic fluid is below normal for the period of gestation, oligohydramnios develops. The possible causes could be decreased production of urine which is caused by bilateral renal agenesis or obstruction of urinary tract and prolonged rupture of the membranes [9] [10].

The classic Potter sequence would occur in fetuses with bilateral renal agenization leading to oligohydramnios. Our newborn's ultrasound showed bilateral polycystic kidney disease and a chest X-ray showed bilateral pulmonary hypoplasia. Therefore, our case appears to be a classic Potter sequence case, a karyotype had done to look for associated genetic or chromosomal abnormalities.

Medical management of newborns with Potter sequence depends on their renal function, respiratory status and associated congenital anomalies [11]. In cases of classic Potter sequence with bilateral renal agenesis, further treatment may not be helpful and the prognosis is serious. However, non-classical Potter sequences due to rupture of membranes during pregnancy have higher chances of survival and require appropriate assessment, resuscitation, and management for better neonatal outcomes.

Our case highlights the importance of regular prenatal checks and examinations in each pregnancy, which helps to collect suspected cases and improve knowledge of this syndrome for better management.

4. Conclusion

Potter sequence is a rare but known complication of chronic oligohydramnios and is associated with a serious fetal prognosis. Prevention and early diagnosis of the underlying cause of oligohydramnios leads to improved neonatal outcomes. However, classic Potter sequence requires early termination of pregnancy, due to underlying complications such as bilateral renal agenesis, unlike cases of non-classical Potter sequence which can usually be prevented and treated to help improve the vital prognosis of newborns.

Disclosures

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